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Bicoronal Synostosis in a Child From Historic Omaha Cemetery 25DK10

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KEY WORDS Omaha Tribe; craniosynostosis; bicoronal fusion

ABSTRACT Fragmentary cranial remains of a child from a commingled burial in a historic Omaha Cemetery (AD 1780–1800) exhibit bony fusion between the frontal, parietals, and sphenoid. The child's remains are consistent with a developmental age between newborn and 6 months postnatal. Radiological and morphological analyses confirm that this individual exhibits osteological signs pathognomic of bicoronal sutural synostosis, including deformation of the lateral orbital margin. This case, although fragmentary, significantly augments other archaeological cases of coronal synostosis reported in the literature. In addition, an extremely large bregmatic fontanel, expanded anterior cranial fossa, and bossed forehead compared with undeformed individuals suggest the child also suffered from increased intracranial pressure perhaps related to an associated hydrocephaly. Despite the deformity, the remains of this child were treated in much the same manner as other infant remains from the site, including the presence of red mercury pigment on the skeletal remains. Am J Phys Anthropol 105:369–376, 1998.

Congenital defects of the skeleton inform us about the health and genetics of past populations as well as about the growth and development of normal and abnormal skeletal elements (e.g., Richards, 1985; Richards and Antón, 1991; Barnes, 1994). Patterns of these defects across skeletal samples allow us to predict the frequencies of more severe congenital conditions in these same samples (Barnes, 1994). Yet the recognition of these patterns relies heavily on the veracity of our diagnostic criteria and the systematic description and publication of individual cases as they are found (Richards and Antón, 1991). Although congenital defects are powerful insights into prehistoric health, the literature on archaeological cases of congenital defects remains relatively sparse (see recent compilations by Barnes, 1994). This is particularly true of cases of premature synostoses of sutures other than the sagittal.

A small number of archaeological cases of coronal synostosis, including both bicoronal and unicoronal fusion, have been reported (Bennett, 1967; Kreiborg and Bjork, 1981; Ortner and Putschar, 1981; Prokopec, 1984; Berrizbeitia, 1992; Webb, 1995). Few of these were extensively analyzed, and all are significantly older than individual 56b (the subject of this study). In addition, all of these are cases of multiple suture fusion, whereas only half of all clinical bicoronal synostoses involve fusion of multiple sutures (Cohen, 1986). The relatively large number of multiple fusions suggests that isolated cases of coronal fusion may go unrecognized or at least unreported. Despite the

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relatively great prevalence of coronal synostoses in modern populations, the relatively small number of reported archaeological cases makes any report of coronal synostosis important.

Clinical and experimental studies indicate a genetic underpinning in most simple synostoses including coronal synostosis (e.g., Cohen, 1986; Mooney et al., 1994, 1996), but environmental factors such as fetal head constraint (Koskinen-Moffett et al., 1982) and artificial cranial deformation (White, 1996) may also influence the frequency of premature sutural fusion.

Coronal synostosis is more common in females than in males (Barnes, 1994) and may appear as an isolated primary fusion or in association with other synostoses or with syndromes such as Apert's and Crouzon's. Patients with uncomplicated coronal synostosis rarely exhibit a mental deficit (Hunter and Rudd, 1977).

Coronal synostosis may be either uni- or bilateral and most often involves not only the coronal suture itself but also the "ring" sutures including the frontosphenoidal, sphenoethmoidal, and occasionally the frontozygomatic (Marsh et al., 1986). Such fusions do not necessarily affect the entire suture but may be restricted to localized foci along the length of the suture. As a result of the restriction of lateral growth, the lateral portion of the orbital plate of the frontal is pulled superiorly relative to the interorbital septum, resulting in an upswept appearance of the lateral orbit on radiographs (Marsh et al., 1986; Cohen, 1986). This orbital deformation is referred to as harlequin eye and is considered pathognomic of coronal synostosis.

The harlequin eye deformation differs from that seen in hydrocephalus in that the medial portion of the superior orbit is pulled upward resulting in a shallow orbital roof ("setting sun sign" [Richards and Antón, 1991]). Individuals with coronal synostosis often exhibit evidence of increased intracranial pressure similar to that seen in hydrocephalus: bulging forehead, depressed orbital plate of the frontal, and silver-beaten appearance of the endocranial vault due to exaggerated sulcal and gyral impressions. In addition, cases of coronal synostosis may also exhibit hypertelorism (Costaras and Pruzansky, 1982).

Here we describe the fragmentary cranial remains of a child discovered in the historic Omaha Tribal cemetery 25DK10. The remains exhibit bilateral fusion among frontal, parietal, and sphenoid bones. Because of their fragmentary nature, developmental age and differential diagnosis of additional syndromic signs are problematical. Following this description, we build on the foundation provided by Richards and Antón (1991) and consider alternative methods for diagnosing pathological conditions.

MATERIALS AND METHODS

Individual 56b was found in a common burial pit with the skeletons of four other children that were recovered in 1989 from a historic Omaha Tribal cemetery 25DK10 by Susan Moorhead and Karl Reinhard (O'Shea and Ludwickson, 1992). Originally dated between 1780 and 1810 (O'Shea and Ludwickson, 1992), touch marks on burial-associated silver artifacts further constrain usage of the cemetery to between 1780 and 1800. Individuals 53 and 54 were fully articulated and died between 6 and 18 months of age, based on radiological assessment of dental development. Individual 55 (fetus) and Individual 56a (neonate) were nearly complete skeletons and skulls. Fragmentary remains from multiple individuals in the same burial are not unusual for this cemetery and probably resulted from the Omaha practice of scaffold burial (O'Shea and Ludwickson, 1992; Reinhard and Ghazi, 1992). As with many other infants in the cemetary, the ectocranial surface of skull of individual 56b was covered with red mercury lead pigment traded to the Omaha and other Missouri River tribes by fur traders (Ghazi et al., 1994; Reinhard and Ghazi, 1992).

The skeletal elements from this multiple burial were examined for additional remains of individual 56b; however, only the frontal bones fused to fragments of the sphenoid and parietals were located. Individual 56b exhibited little evidence of weathering, distortion, or mineral staining (Figs. 1–3). The specimen consists of left and right frontal bones that do not meet at the midline but are fused bilaterally to the parietal bones

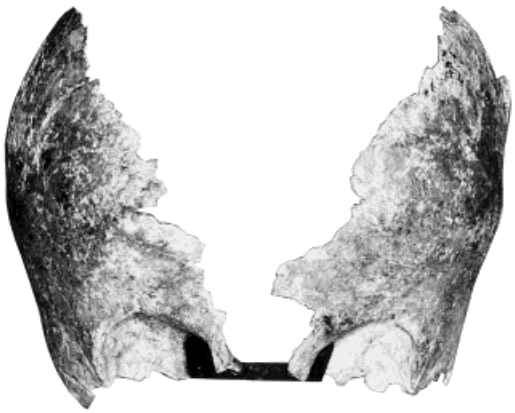

Fig. 1. Remains of individual 56b arranged in anatomical association (ectocranial view).

and greater and lesser wings of the sphenoid along the coronal and frontosphenoidal sutures. Only small portions of the parietal bones and greater wings of the sphenoid remain on the lateral vault wall.

Because of the fragmentary nature of the specimen, we had neither standard data points nor reference planes that would have enabled us to utilize normative databases for comparison. Even so, specimens this young are underrepresented in the literature, and the potential comparative databases for such things as biorbital breadth don't have individuals that young for comparison (Richards, 1985; Richards and Antón, 1991). Therefore, we had to compare our individual with the skeletal remains of same-aged individuals with similar conditions using the same measurements in the same planes as were available on our specimen.

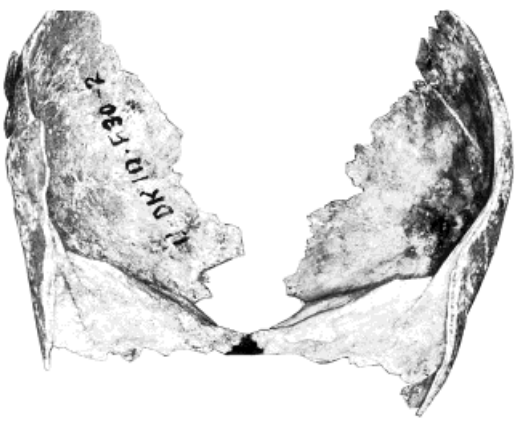

Fig. 2. Remains of individual 56b arranged in anatomical association (endocranial view).

Fig. 3. Remains of individual 56b arranged in anatomical association (lateral view).

Individual 56b was radiographed, reconstructed, and measured by one of us (S.C.P.) and visually examined by both of us to determine developmental age at death and the degree and kind of pathological conditions present. Individual 56b was reconstructed on the basis of the interorbital width and the relationship between the superior margin of the superior orbital fissure, the frontoethmoidal suture, and the width of the cribriform plate as assessed relative to the comparative sample (Fig. 4). The comparative sample included recent undeformed skulls (n = 5; neonate, 3, 4, and 11 years, and adult), an immature hydrocephalic from the Dental Historical Museum, University of Nebraska Medical Center (UNMC), and three undeformed and one hydrocephalic neonate from the Atkinson Collection, University of the Pacific School of Dentistry (UOP). In addition to the skulls, lateral cephalograms of children 3–8 months of age (n = 5) from the Colorado Study Database, Department of Pedodontics, UNMC, were used for comparative purposes.

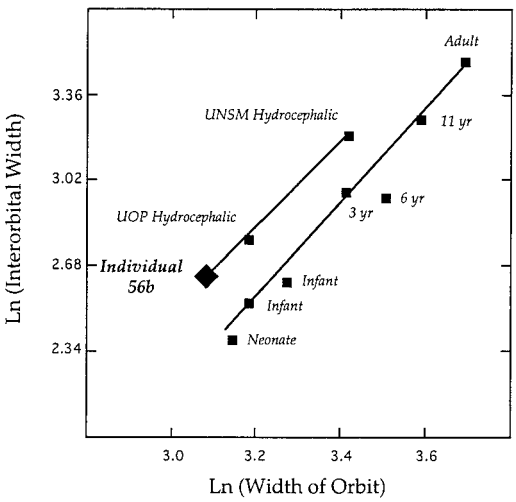

Fig. 4. Scatter plot of Ln (interorbital width) v. Ln (orbital width).

The preservation of individual 56b limited the measurements to interorbital width, internal biorbital width, and width of the orbit defined as the contribution of the frontal bone to the orbital rim. These measurements were taken on skulls in the comparative sample (Table 1). Bivariate plots of log-transformed data of orbital vs. interorbital width are used to assess the relative development of the interorbital region. Least-squares regressions were calculated by group (Table 2).

Each skull was radiographed in sagittal, anteroposterior, and superior views using standard orientation procedures. Radiographs were taken at the Health Center (University of Nebraska, Lincoln) and at UNMC. Two angles were measured on each radiograph: 1) in superior view, the posteriorly directed angle between the lesser wings of the sphenoid was measured along the posteriormost margin of the anterior cranial fossa, and 2) in anterior view, the superiorly directed angle formed between the lesser wings of the sphenoid was measured along the superior endocranial surface adjacent to the frontosphenoidal sutures. Bivariate plots of these angles (Fig. 5) illustrate the relative development of the lesser wings of the sphenoid.

TABLE 1. Measurements (mm)[1]

Specimen	IOW	BOW	OW	SUP	ANT
Neonate	11.2	—	23.0	—	—
Infant	12.7	—	24.0	—	—
Infant	13.8	—	26.3	—	—
3 years	19.7	80.2	30.2	101	120
6 years	19.2	85.4	33.1	108	130
11 years	26.3	98.3	36.0	113	135
Adult	33.0	112.8	39.9	128	142
Individual 56b	14.0	53.4	22.0	—	—
UOP hydrocephalic	16.5	—	24.0	—	—
UNSM hydrocephalic	24.7	85.5	30.4	125	125

[1]ANT, angle between lesser wings of sphenoid bone viewed anteriorly; BOW, biorbital width; IOW, interorbital width; OW, width of single orbit; SUP, angle between lesser wings of sphenoid bone viewed superiorly.

Sagittal and anteroposterior radiographs of each specimen were traced onto acetate sheets. The selection of registration points was limited by the preservation of individual 56b. Tracings of radiographs of each comparative skull were overlain simultaneously upon the frontoethmoidal sutural margin (Fig. 6). Tracings of the known hydrocephalic from UNMC and individual 56b were then registered upon the tracings of undeformed skulls. A similar process was employed with sagittal tracings to evaluate the shape of the frontal bones and the relative size of the bregmatic fontanel (Figs. 6, 7).

RESULTS

Description

Based on extrapolation from Figures 4 and 5 as well as the relative size and morphological development of individual 56b, we determined that the child was possibly a neonate but no more than 6 months of age at the time of death.

In frontal view, individual 56b presents two fairly complete frontal bones that do not meet at the midline along the frontal squama. Close examination of the free medial edges of each frontal bone, particularly in the region of the anterior fontanel, show that the bone was not broken postmortem but tapers out, suggesting a large, patent fontanel. The absence of the parietal precludes a definitive statement on the sagittal extent of the fontanel. However, the anterior portion of the fontanel between the frontal bones is estimated to be 70 mm long and 60 mm wide. The orbital plates of the frontal are depressed relative to the frontal squama, resulting in a relatively shallow superior

TABLE 2. Regression analysis

Ln (IOW) vs. Ln (OW)	Combined (n = 10)	Normal (n = 7)	Deformed (n = 3)
Slope	1.57 ± 0.246	1.86 ± 0.151	1.74 ± 0.039
Y-intercept	−2.36 ± 0.825	−3.43 ± 0.513	−2.75 ± 0.124
Homogeneity of slopes		$P = 0.065$	ns
Homogeneity of intercepts		$P = 0.033$	*

* Significant.

orbit. In addition, the lateral edge of each orbit appears swept up relative to the interorbital septum. This appearance is confirmed by the radiographic examination (see below).

Laterally, dense lamellar bone occludes the coronal suture between the sphenoid and parietal. This fusion does not extend the entire length of the coronal suture but is localized to its more inferior portions. The frontosphenoidal suture within the anterior cranial fossa is contiguous with the coronal suture and exhibits bilateral fusion as well.

The frontal bones are smooth on their ectocranial surfaces without any marked indentations. The sagittal profile of the frontal bones suggests that they are strongly bowed anteriorly, suggesting slightly increased intracranial pressure. Endocranially, the sulcal and gyral impressions of individual 56b are unremarkable, suggesting no marked increase in intracranial pressure.

Metric results

Bivariate plots show that both modern hydrocephalics and individual 56b exhibit somewhat wider interorbital widths than do the comparative skulls (Fig. 4). Whereas pathological and nonpathological regression slopes do not differ from each other, y-intercept values are significantly different from each other (Wilkinson, 1990) (Table 2). Orbital height could not be assessed due to the incomplete nature of individual 56b.

Bivariate plots suggest expansion of the anterior cranial fossae in both the UNMC hydrocephalic and individual 56b (Figs. 4, 5); both have relatively broad superior sphenoidal angles compared with the size of their anterior sphenoidal angles. The UOP hydrocephalic was not measured. The UNMC hydrocephalic exhibits more extreme broadening of the superior sphenoid angle, suggesting more extreme rotation of the sphenoid anteriorly and laterally. Individual 56b exhibits upswept lateral orbits relative to AP radiographs of the comparative sample (Fig. 6).

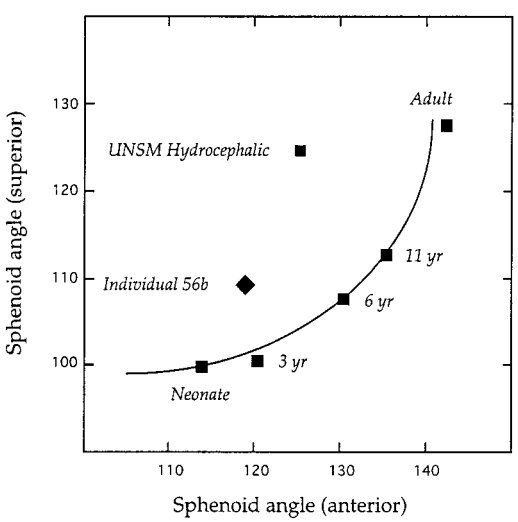

Fig. 5. Bivariate plot of the angle of the sphenoid angle (viewed superiorly) vs. the sphenoid angle (viewed anteriorly).

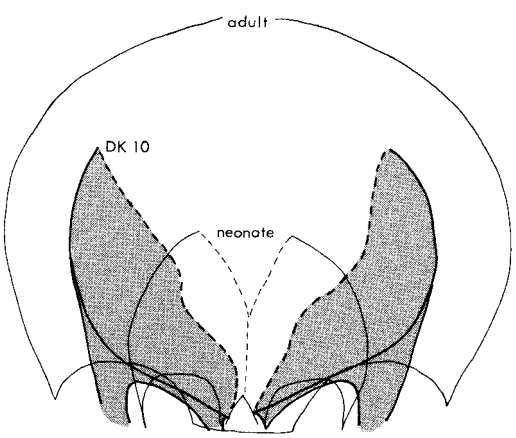

Fig. 6. Registration of tracings of the frontal bones (viewed anteriorly) of individual 56b and those of normal adult and neonate tracings.

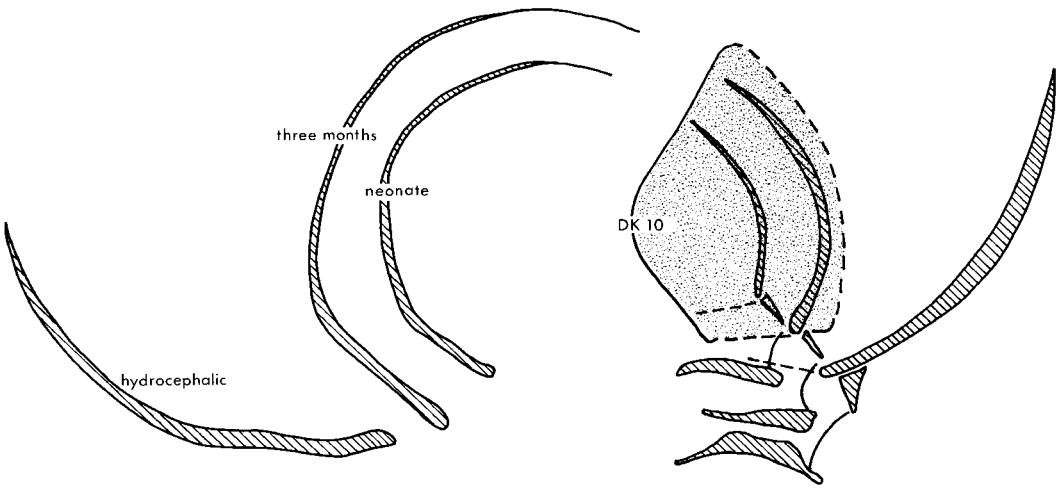

Fig. 7. Registration of tracings of the frontal bones (viewed laterally) of individual 56b and those of normal adult and neonate tracings.

Both the UNMC hydrocephalic and individual 56b exhibit bossed frontals when compared with the lateral cephalograms of 3-month-old and neonate individuals (Fig. 7). In this figure, the anteriormost edge of individual 56b's tracing is dotted to depict the osteological limit of the frontal bone where it borders the frontal fontanel. The actual curvature of individual 56b's bossed frontal would have extended well beyond this dotted line and beyond the 3-month-old profile.

DISCUSSION AND DIFFERENTIAL DIAGNOSIS

Individual 56b represents the remains of a young infant with bony fusion between frontal, sphenoid, and parietals. In conjunction with the upsweeping of the lateral orbit, a pathognomic indicator of coronal synostosis, this fusion leaves no doubt that bicoronal synostosis must form part of the diagnosis for this individual. The increased interorbital breadth relative to orbital breadth is also consistent with bicoronal synostosis (Costaras and Pruzansky, 1982). However, the fragmentary remains of this individual make a differential diagnosis and the assessment of any complicating factors problematical.

The greatly enlarged bregmatic fontanel of individual 56b, in conjunction with the results of the present cephalometric analysis, indicates that this infant exhibited a moderately bossed forehead and enlarged anterior cranial fossa. Together, these observations suggest that Individual 56b may also have had increased intracranial pressure related to mild hydrocephalus (Richards and Antón, 1991). If the hydrocephalus were secondary to the synostosis, the tendency toward patent lateral fontanels seen in many hydrocephalics would not be exhibited in this individual and might be compensated for by enlargement of the bregmatic fontanel. However, the definitive indicator of increased intracranial pressure, the silver-beaten appearance of the endocranial surface, is not in evidence. Other indicators of hydrocephalus, including increases in endocranial volume, relative proportions of the vault, and appearance of the dural sinuses and sutures, cannot be assessed in individual 56b due to its fragmentary nature. Although the medial superior orbit is relatively shallow, it is not clear whether this appearance is related predominantly to deformation of the lateral orbit caused by the coronal synostosis or is a result of medial orbital shallowing often seen in hydrocephalus (e.g., Richards and Antón, 1991). Thus, although an association with mild hydrocephalus is suggested, it cannot be a certainty.

Although some syndromatic diagnoses such as achondroplasia can be ruled out on the basis of individual 56b's highly enlarged bregmatic fontanel and lack of deformation around the metopic suture, other syndromes cannot be ruled out. Amongst the syndromes that might be consistent with the appearance of individual 56b are Apert's syndrome (Cohen and Kreiborg, 1990) and Cloverleaf skulls (Cohen, 1986, 1988). Apert skulls exhibit premature fusion of the coronal and frontosphenoidal sutures and large, patent anterior and posterior fontanels (Cohen and Kreiborg, 1990; Kreiborg and Cohen, 1990). In isolation, the frontal bones of Cloverleaf skulls are not unlike those in Apert's syndrome. However, the remains of individual 56b are too fragmentary to make a definitive statement with regard to these syndromes.

The description of this case of bicoronal synostosis in individual 56b increases the relatively small number of archaeological cases of bicoronal synostosis. Individual 56b is the youngest of these reported cases. Individual 56b joins three other cases of sutural synostoses in Omaha cemeteries: two individuals with sagittal synostosis and one with squamosal synostosis (Owsley et al., 1994). In addition to sutural synostoses, other minor congenital conditions including brachydactyly, supernumerary teeth, and atlantooccipital fusion were also present in these cemeteries. Numerous other cases of noncongenital childhood disease and stress have been reported for the Omaha (O'Shea and Ludwickson, 1992; Reinhard and Ghazi, 1992; Reinhard et al., 1994; Ghazi et al., 1994; Owsley et al. 1994; Sandness and Green, 1994). Given the fragmentary nature of the remains, it is not possible to know to what extent the child appeared physically or behaviorally different from other children. Despite the deformity, the remains of this child were treated in much the same manner as other infants from the cemetery, including the presence of red mercury pigment on their skeletal remains. These studies, in conjunction with analyses of adult remains, have resulted in a wealth of knowledge about the health of the Omaha in historic times.

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